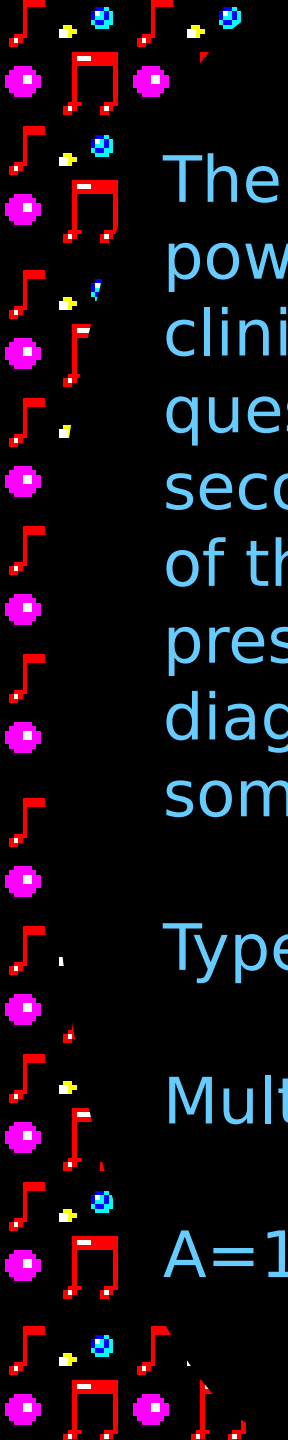


Pathology of the Ear and Temporal Bone

Carol Adair, MD

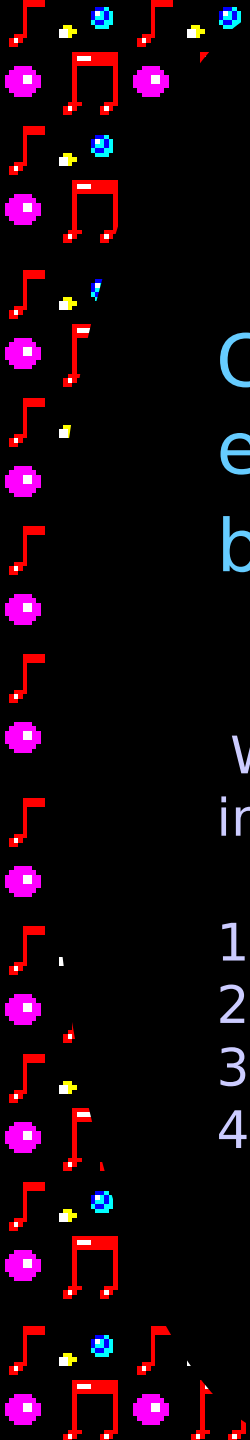


The following series of cases is presented in power point format. The first slide gives the clinical history and a multiple choice (type K) question (see below for answer options). The second and third slides are microscopic images of the lesion in question. The fourth slide presents the answer to the question. The diagnosis is given on the fifth slide, along with some key points.

Type K Question Format:

Multiple Choice:

A=1,2,3 B=1,3 C=2,4 D=4 E=all

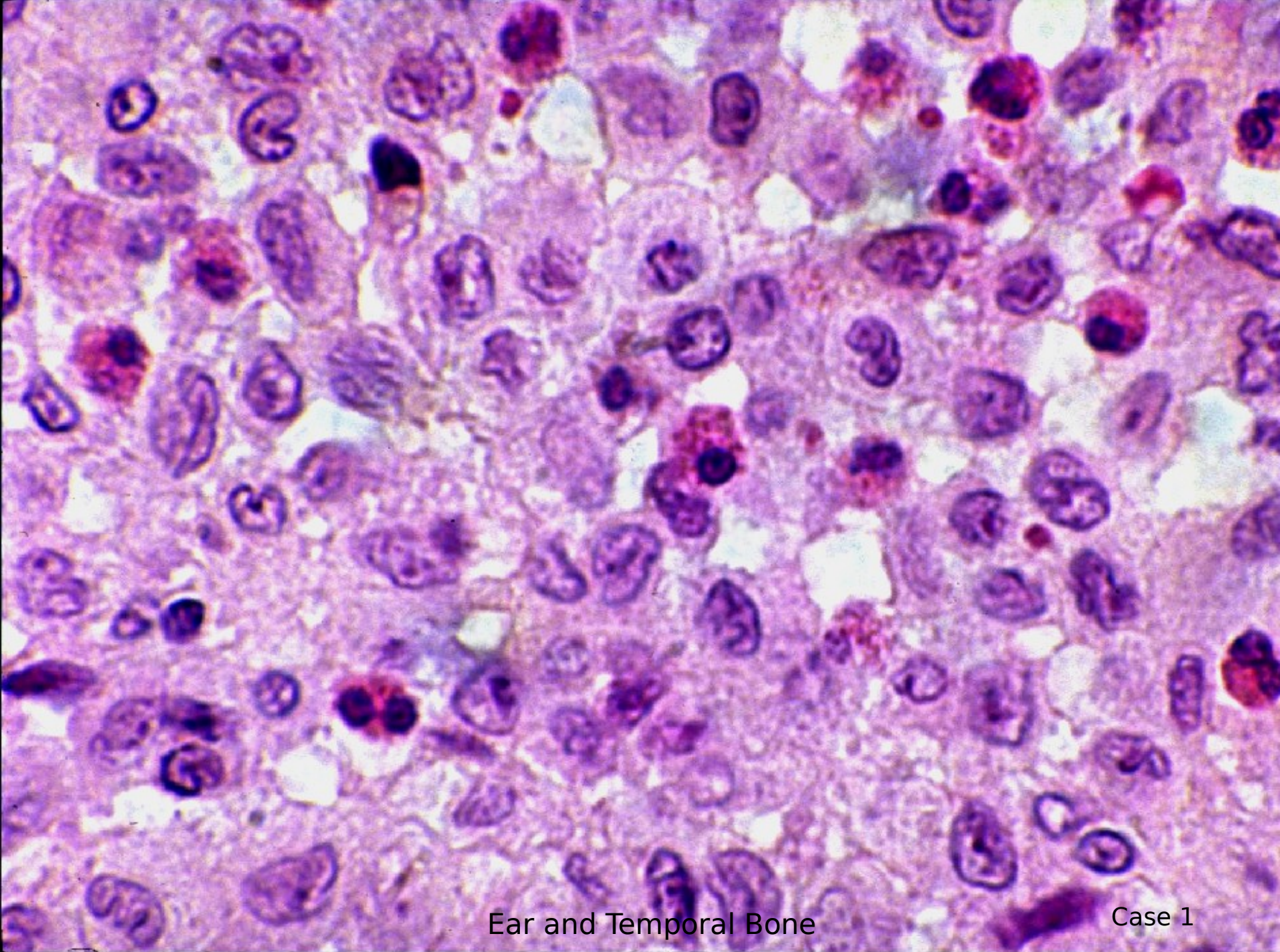


Case 1: A four year-old boy has an expansile lytic lesion of the temporal bone.

Which of the following pathologic features is often seen in this lesion?

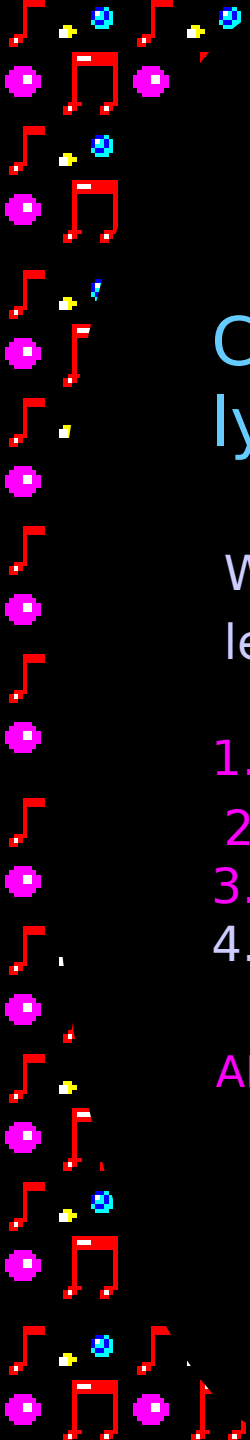
1. Positive for CD 1a by immunohistochemistry.
2. Positive for S100 protein by immunohistochemistry.
3. Birbeck granules by electron microscopy.
4. Premelanosomes by electron microscopy.





Ear and Temporal Bone

Case 1

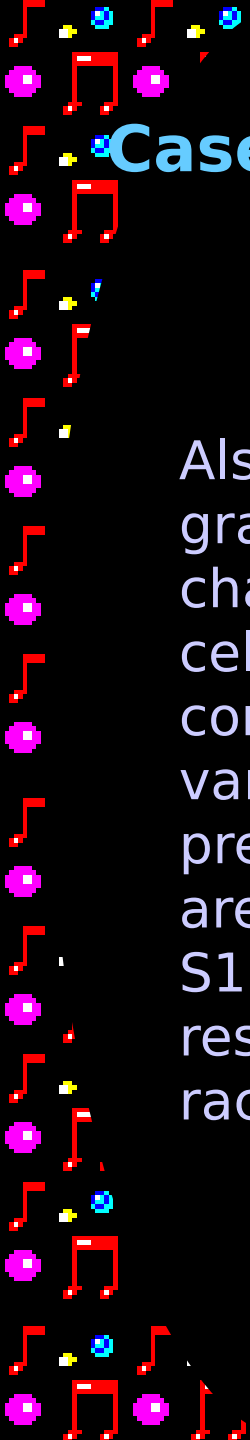


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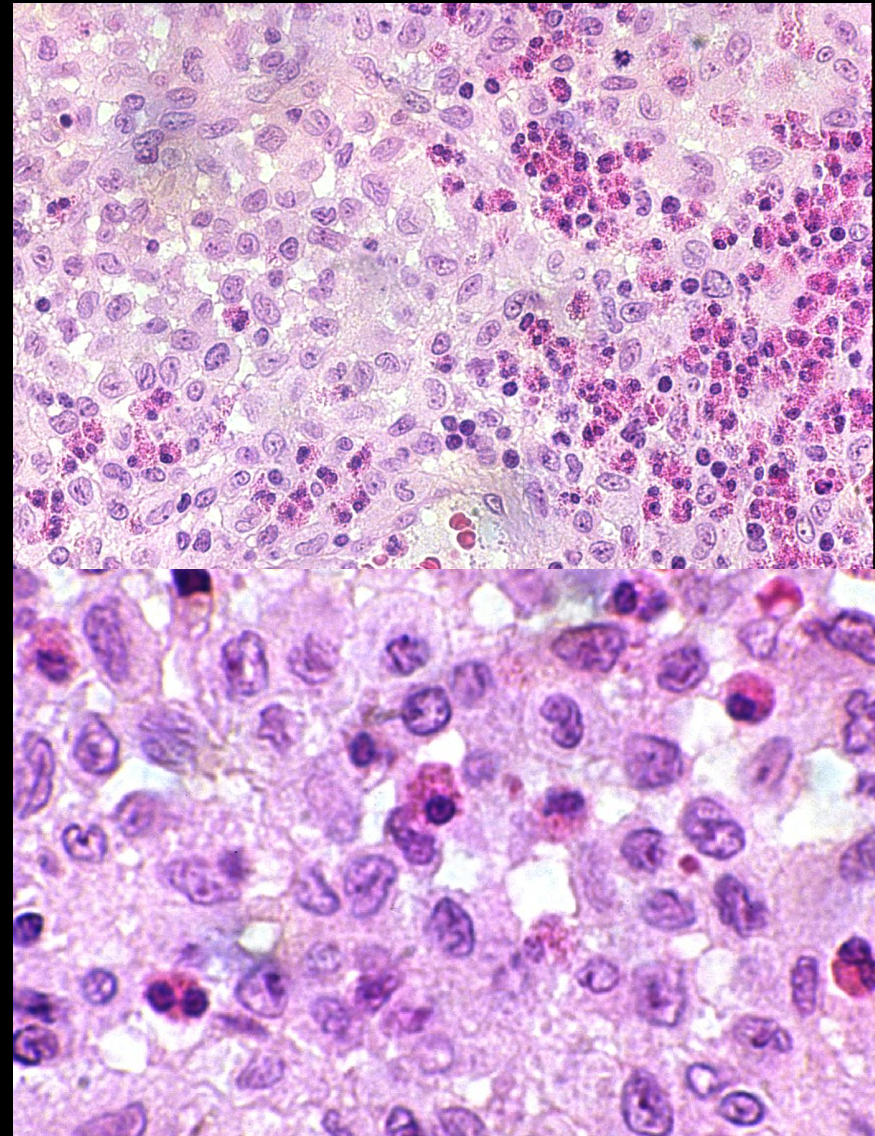
1. Positive for CD 1a by immunohistochemistry.
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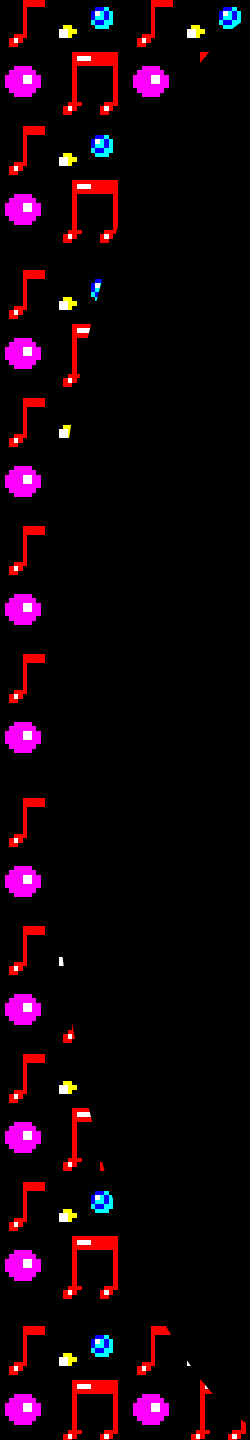
ANSWER: A



Case 1: LANGERHANS CELL HISTIOCYTOSIS

Also known as eosinophilic granuloma, the lesion is characterized by histiocytic cells with grooved or convoluted nuclei. Eosinophils vary in number but are usually present. The histiocytic cells are positive for CD1a and S100. Birbeck granules resemble “zippers” or “tennis rackets” on EM.

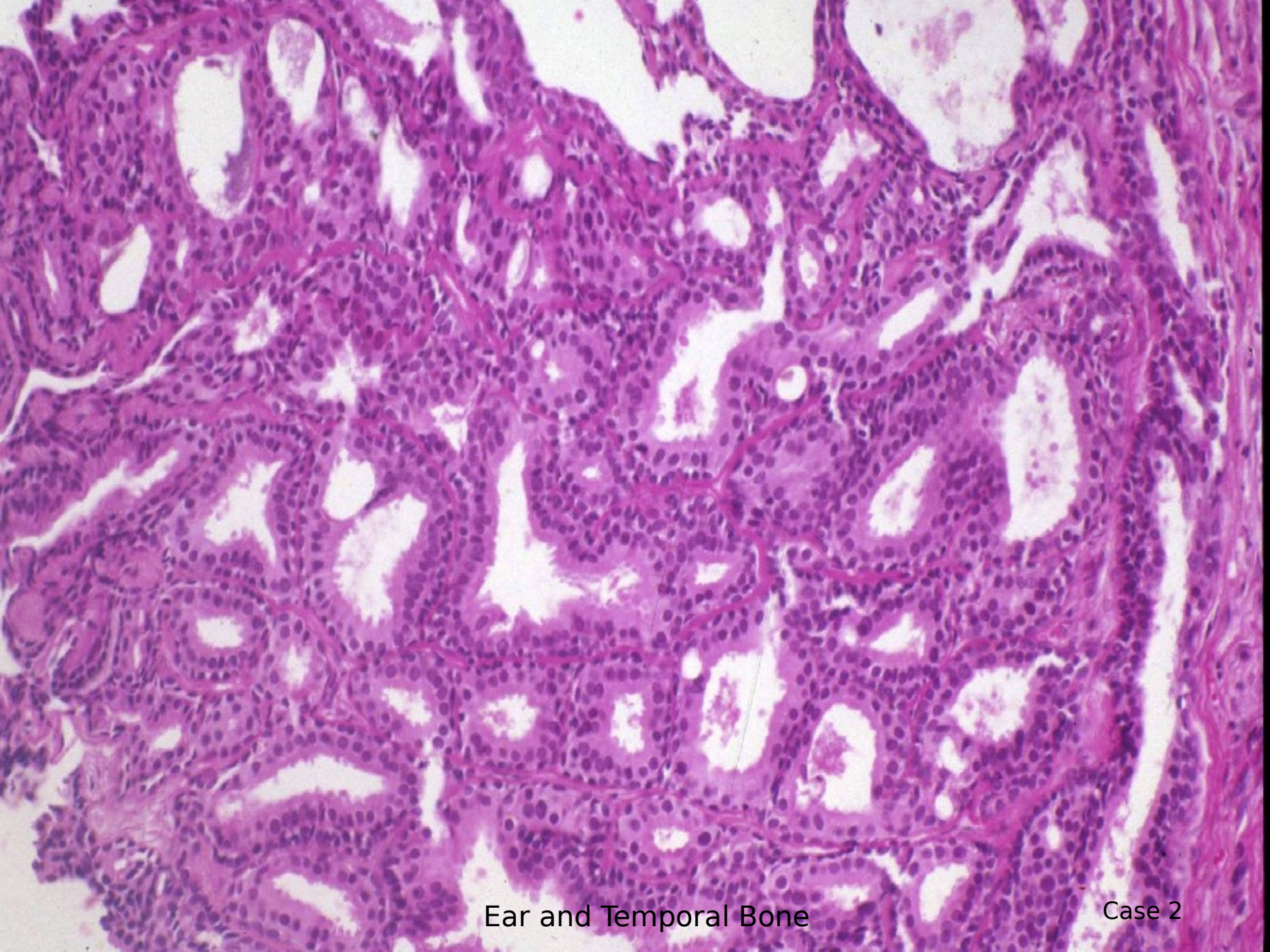


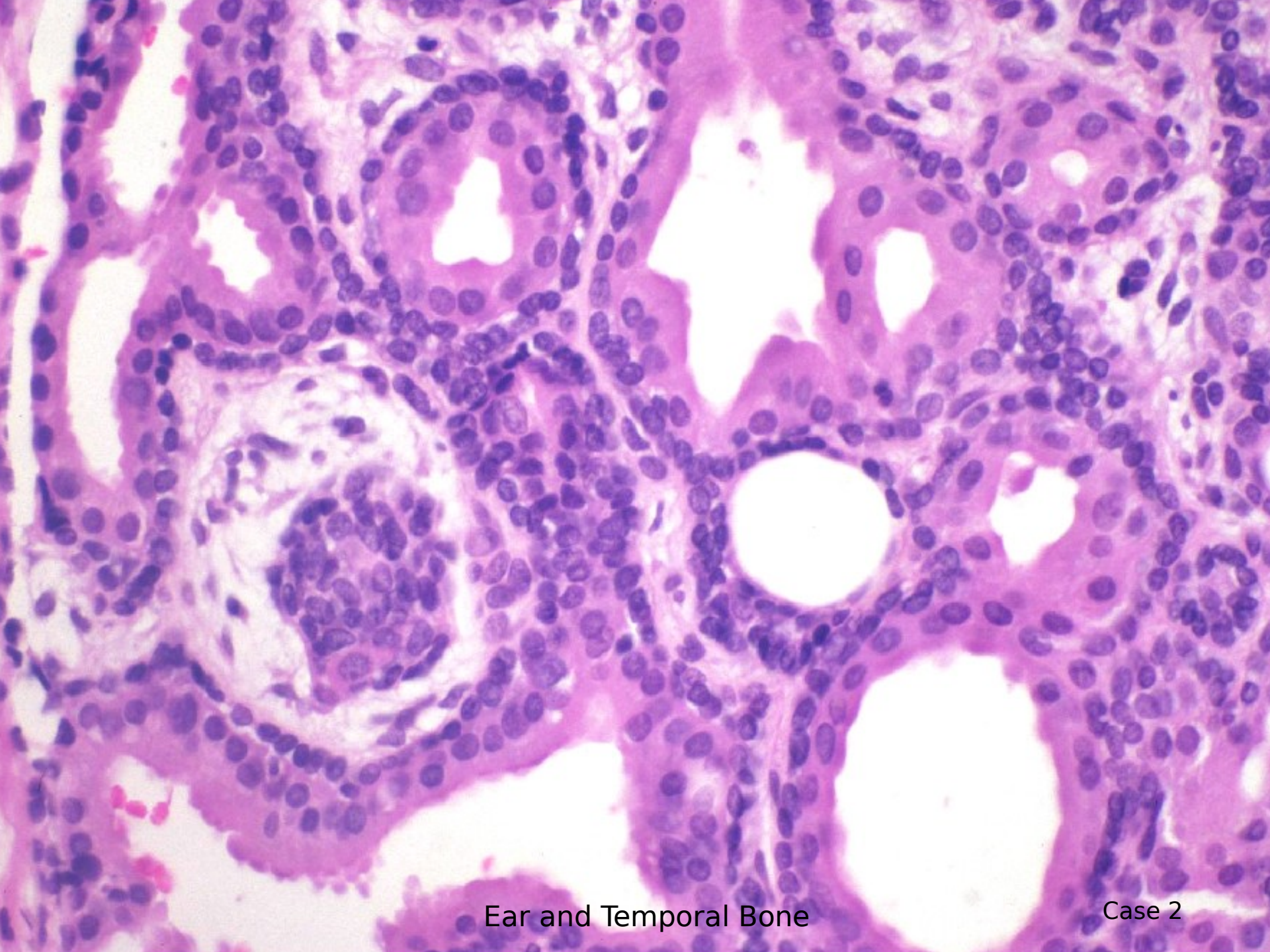


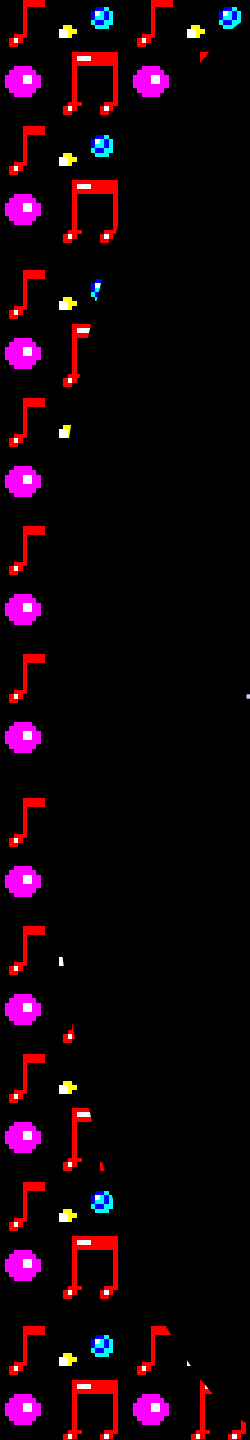
Case 2: A 50 year old male has decreased auditory acuity in the left ear. A nodular lesion is seen elevating the mucosa of the external auditory canal.

The following tumors may arise from the ceruminous glands of the external auditory canal:

1. Ceruminous gland adenoma.
2. Mixed tumor.
3. Adenoid cystic carcinoma.
4. Syringocystadenoma papilliferum.







Case 2: A 50 year old male has decreased auditory acuity in the left ear. A nodular lesion is seen elevating the mucosa of the external auditory canal.

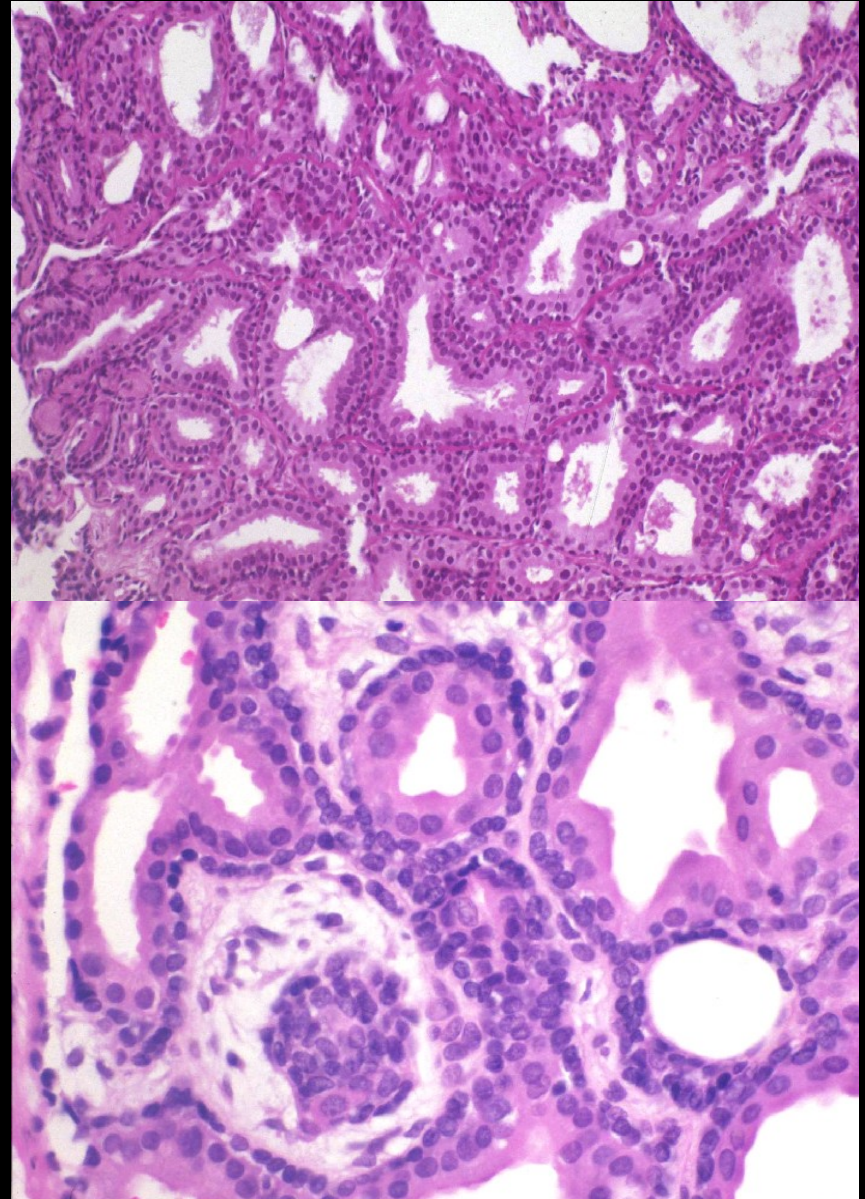
The following tumors may arise from the ceruminous glands of the external auditory canal:

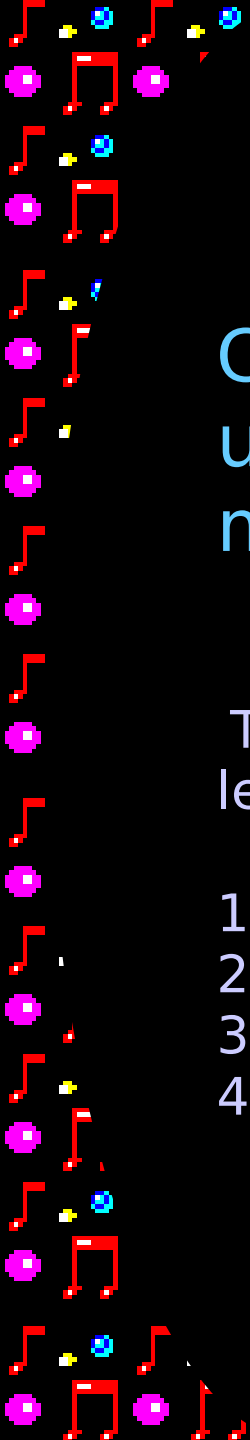
1. Ceruminous gland adenoma.
2. Mixed tumor.
3. Adenoid cystic carcinoma.
4. Syringocystadenoma papilliferum.

ANSWER: E

Case 2: CERUMINOUS GLAND ADENOMA

The ceruminous neoplasms include ceruminous gland adenoma, characterized by apocrine type differentiation, as well as salivary gland type neoplasms, especially mixed tumors and adenoid cystic carcinomas. Syringocystadenoma is another apocrine type neoplasm which arises from these glands.

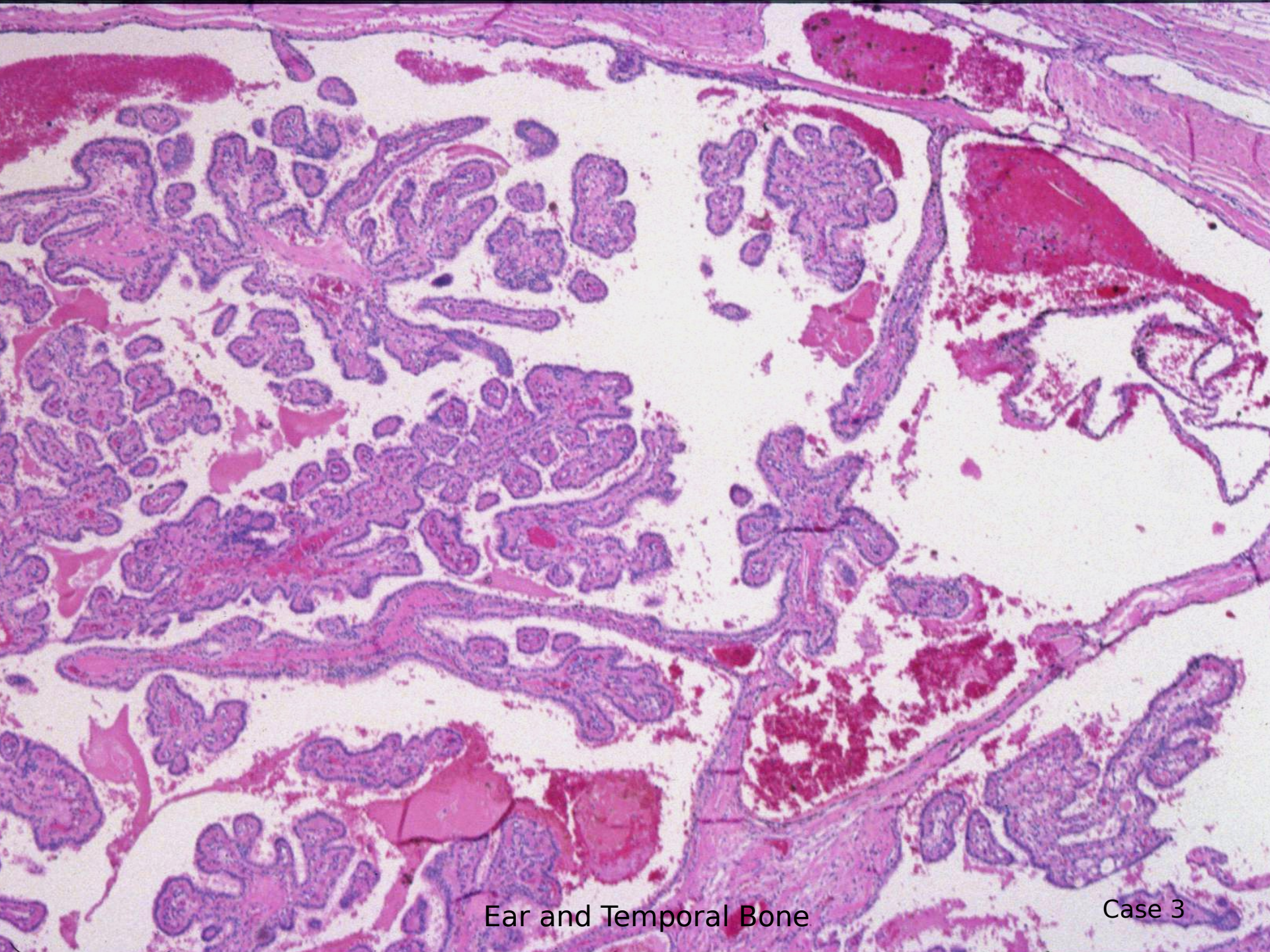




Case 3: A 28 year old female has unilateral hearing loss and headaches. A mass lesion is seen in the temporal bone.

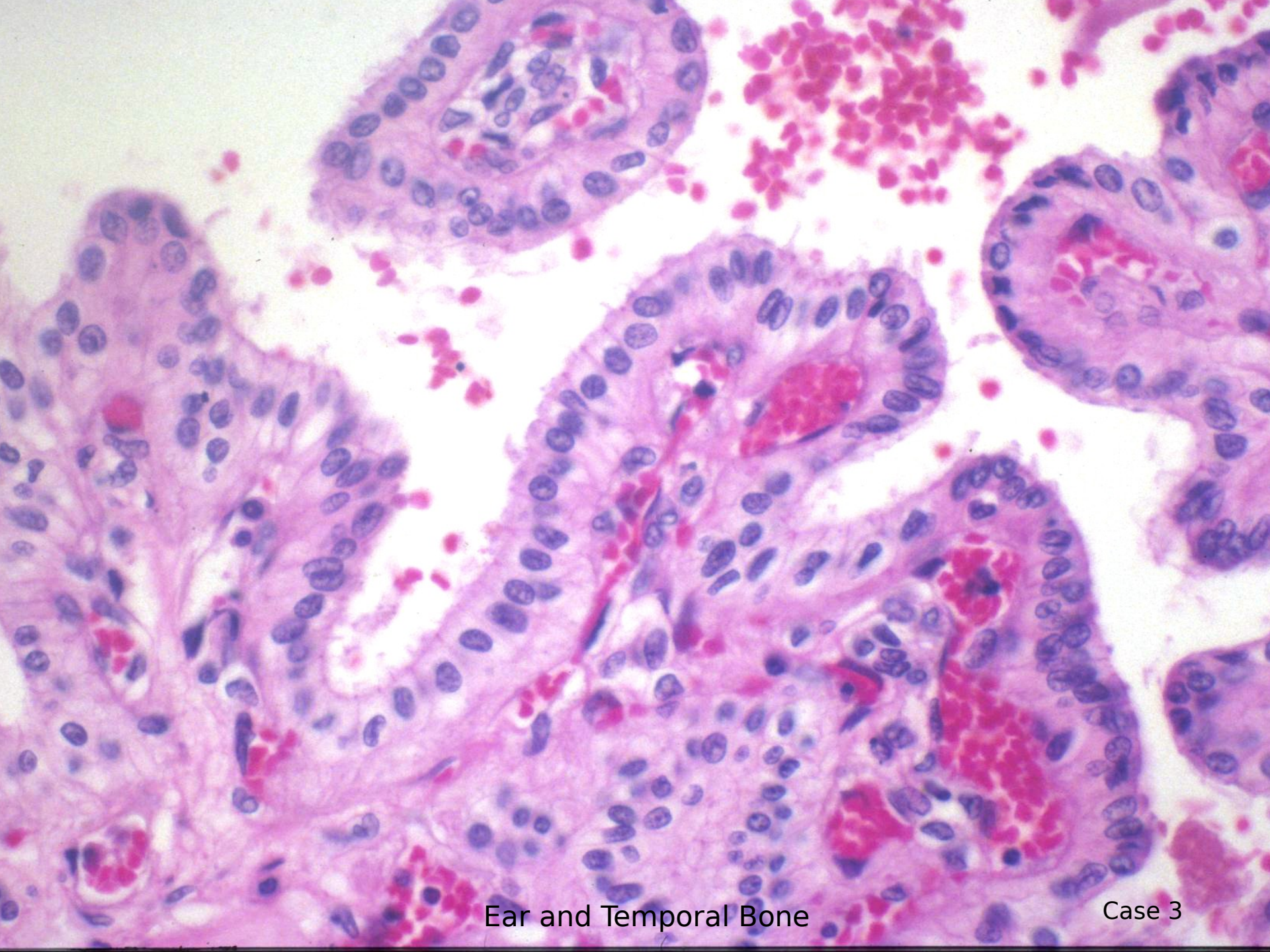
The following syndromes have been associated with this lesion:

1. MEN 2b.
2. Tuberous sclerosis
3. Neurofibromatosis.
4. Von Hippel Lindau disease.



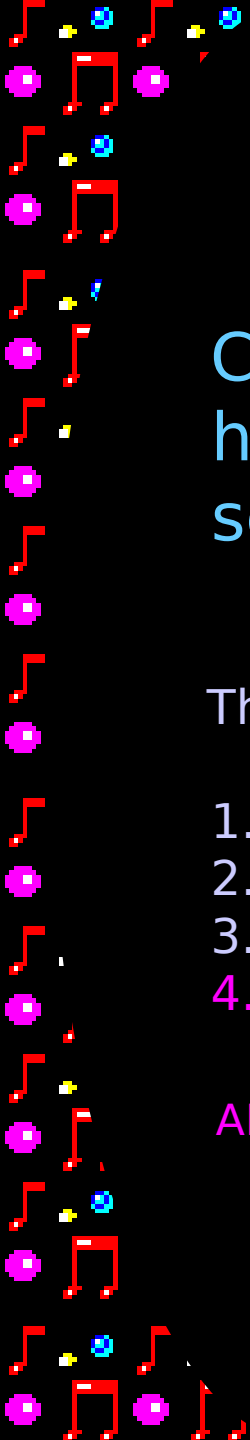
Ear and Temporal Bone

Case 3



Ear and Temporal Bone

Case 3



Case 3: A 28 year old female has unilateral hearing loss and headaches. A mass lesion is seen in the temporal bone.

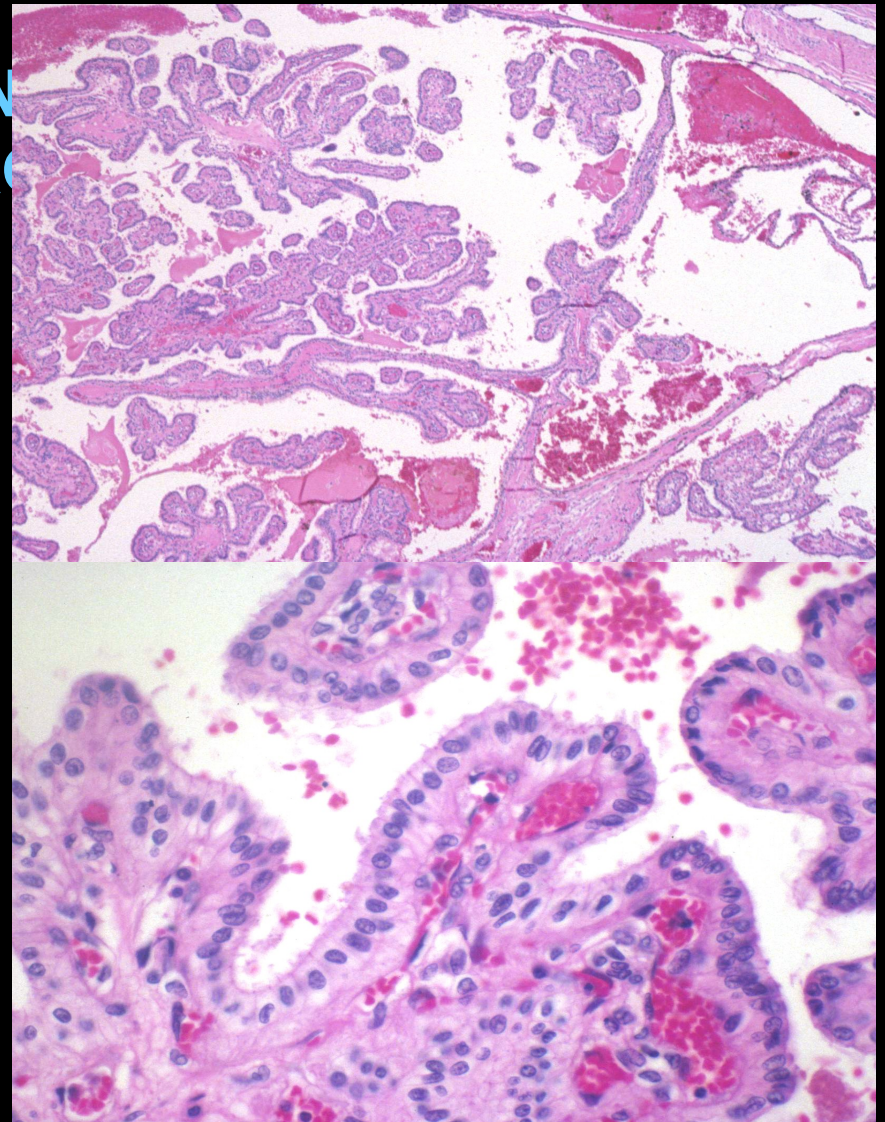
The following syndromes have been associated with this lesion:

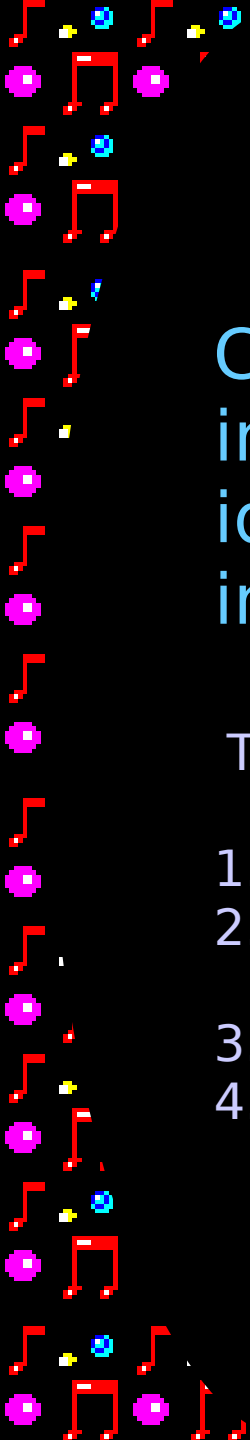
1. MEN 2b.
2. Tuberous sclerosis
3. Neurofibromatosis.
4. Von Hippel Lindau disease.

ANSWER: D

Case 3: PAPILLARY M ENDOLYMPHATIC SAC

Endolymphatic sac tumors are benign but locally destructive lesions which should be treated aggressively; failure to do so may result in death due to intracranial extension. These tumors may be sporadic or may be seen in patients with von Hippel Lindau disease

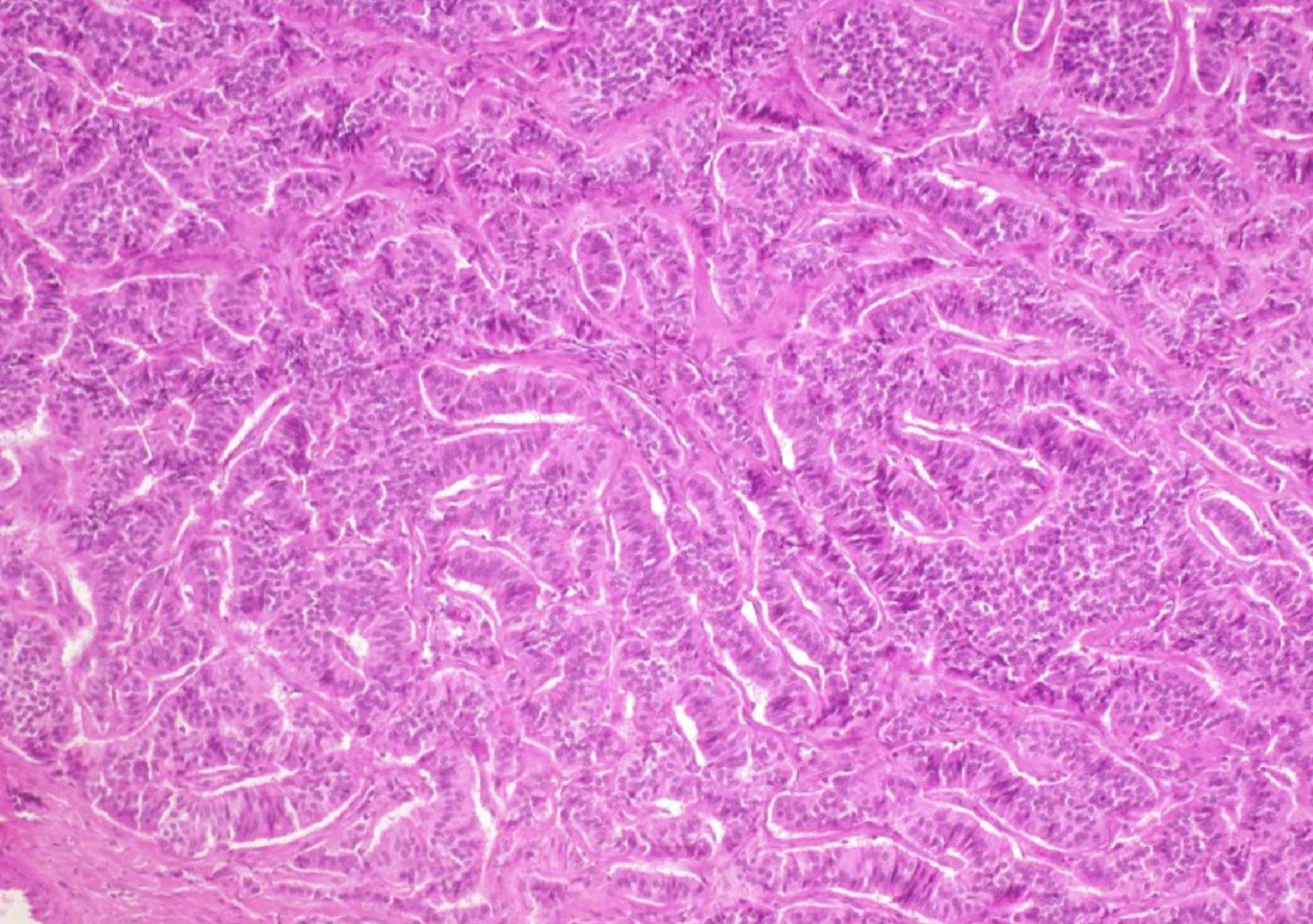


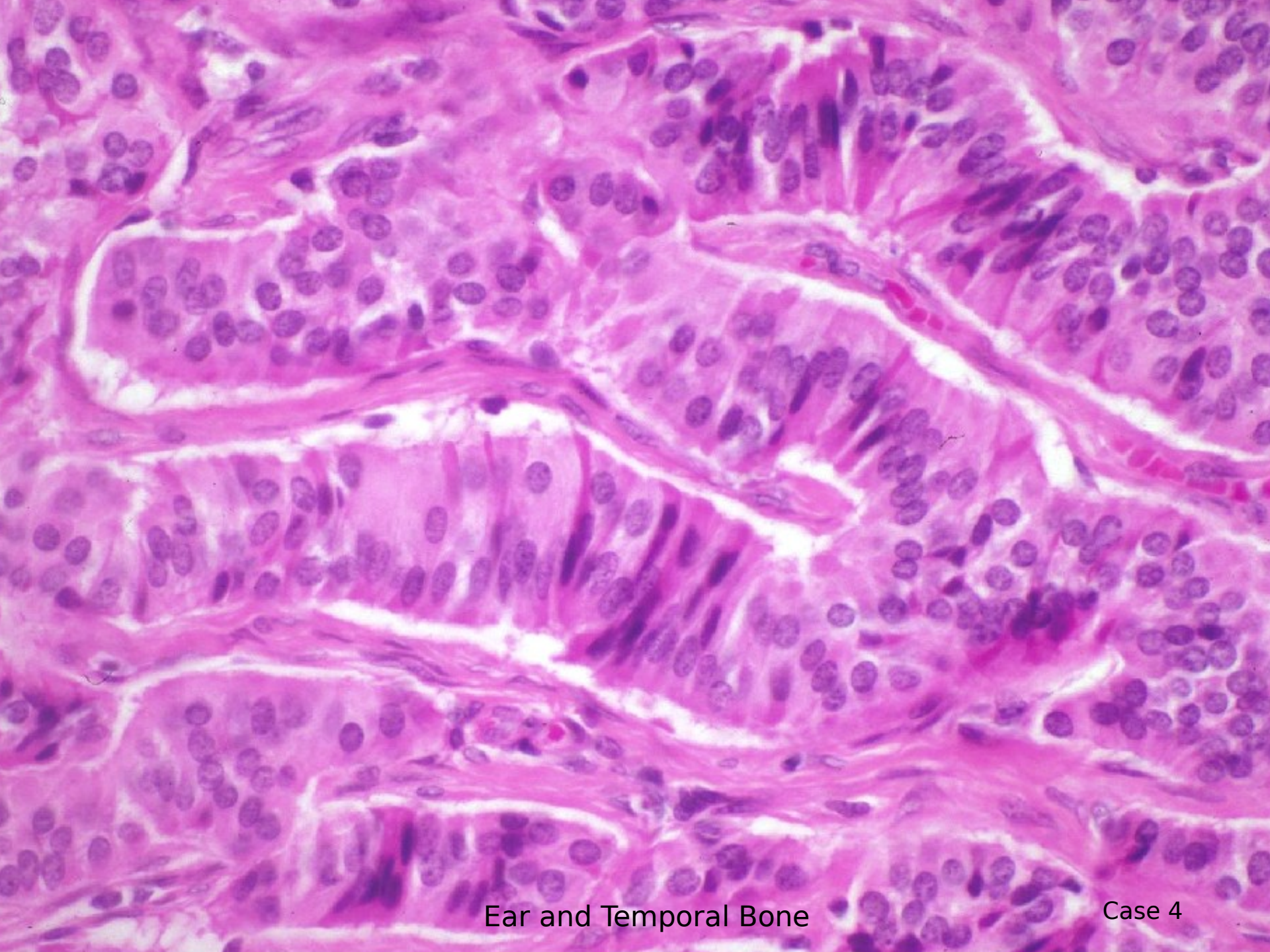


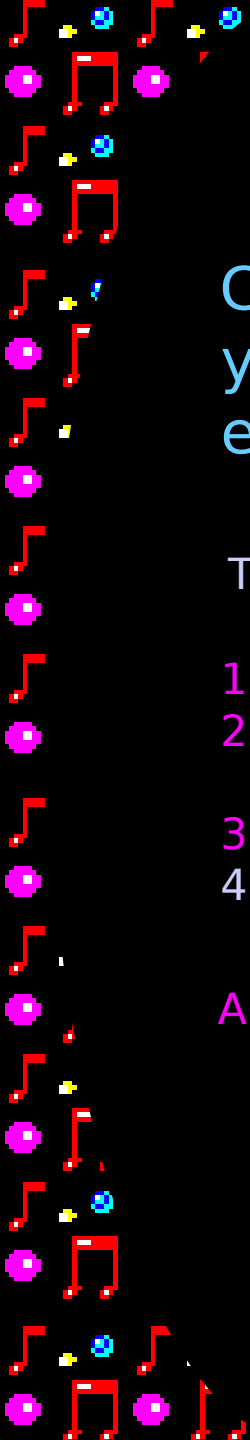
Case 4: Decreased auditory acuity is noted in a 40 year old female. A mass is identified in the middle ear space, with an intact tympanic membrane

The following statements are true of this lesion:

1. Arise from the middle ear mucosa.
2. Is highly vascular by radiologic imaging, making making distinction from paraganglioma difficult.
3. May be positive for neuroendocrine markers.
4. Distant metastasis occurs in 20% of cases.







Case 4: Decreased auditory acuity is noted in a 40 year old female. A mass is identified in the middle ear space, with an intact tympanic membrane

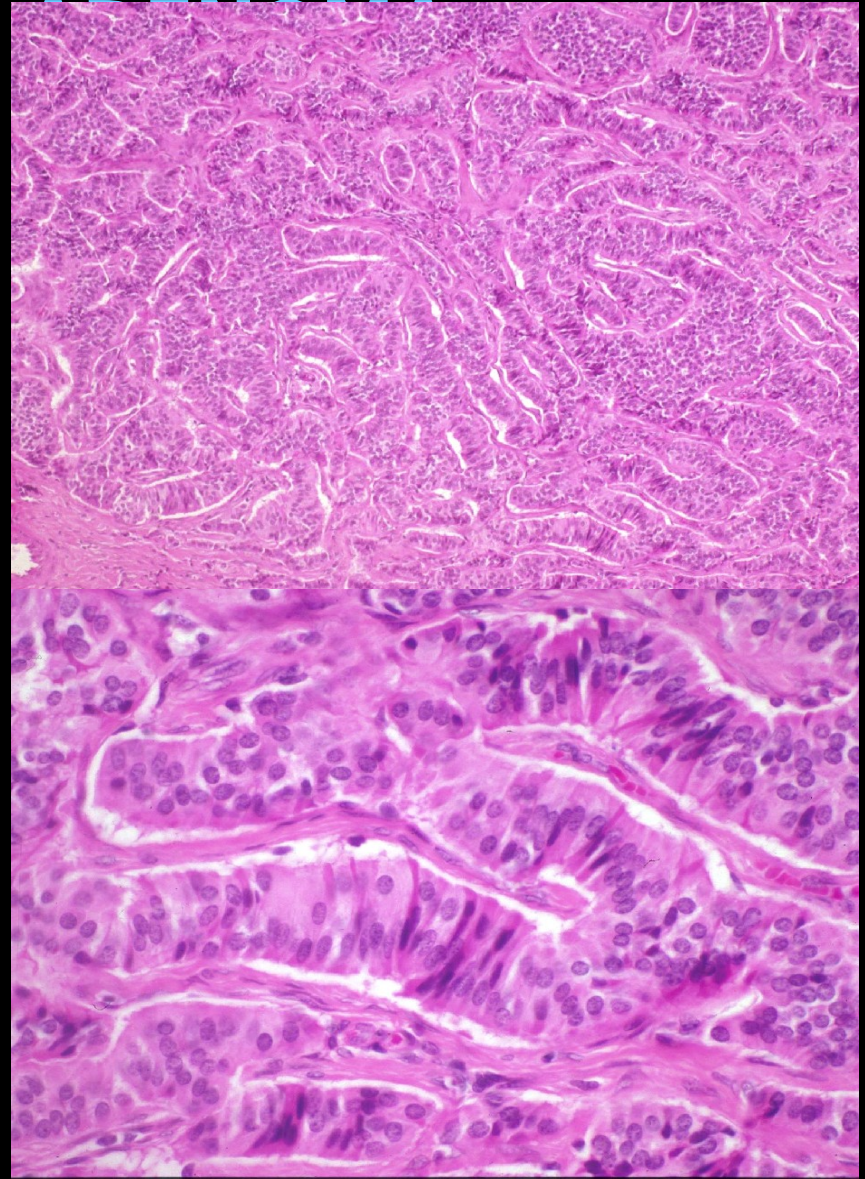
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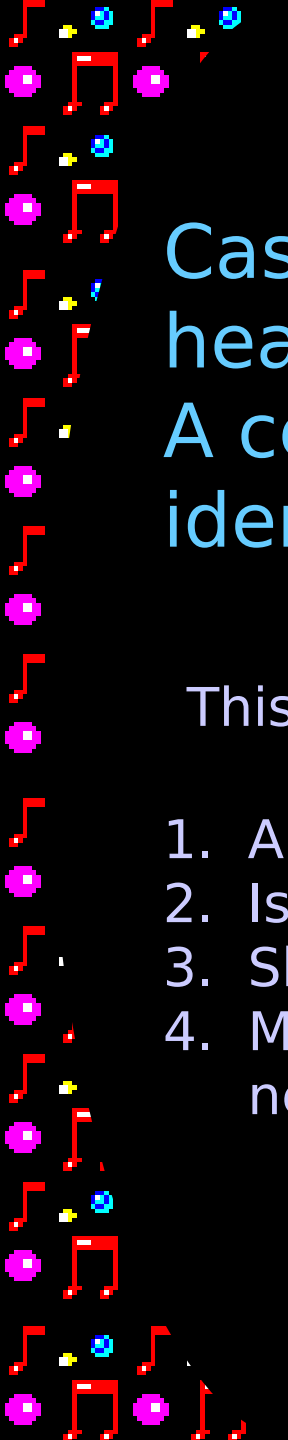
1. Arise from the middle ear mucosa.
2. Is highly vascular by radiologic imaging, making making distinction from paraganglioma difficult.
3. May be positive for neuroendocrine markers.
4. Distant metastasis occurs in 20% of cases.

Answer: B

Case 4: MIDDLE EAR

Middle ear adenomas are benign lesions. They may be locally destructive, but have not been shown to metastasize. Some of these tumors demonstrate neuroendocrine markers (carcinoid-like) and some do not.

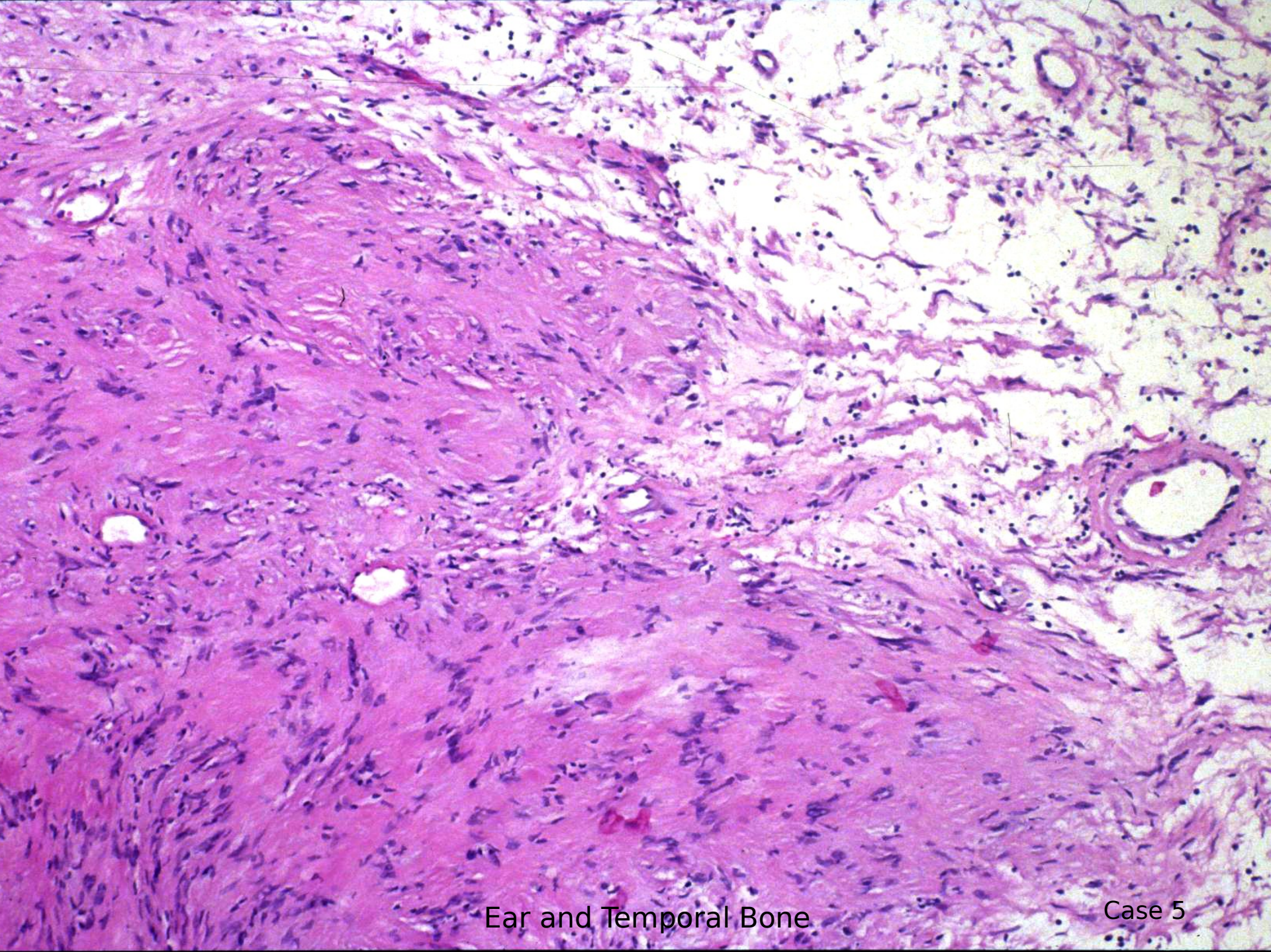


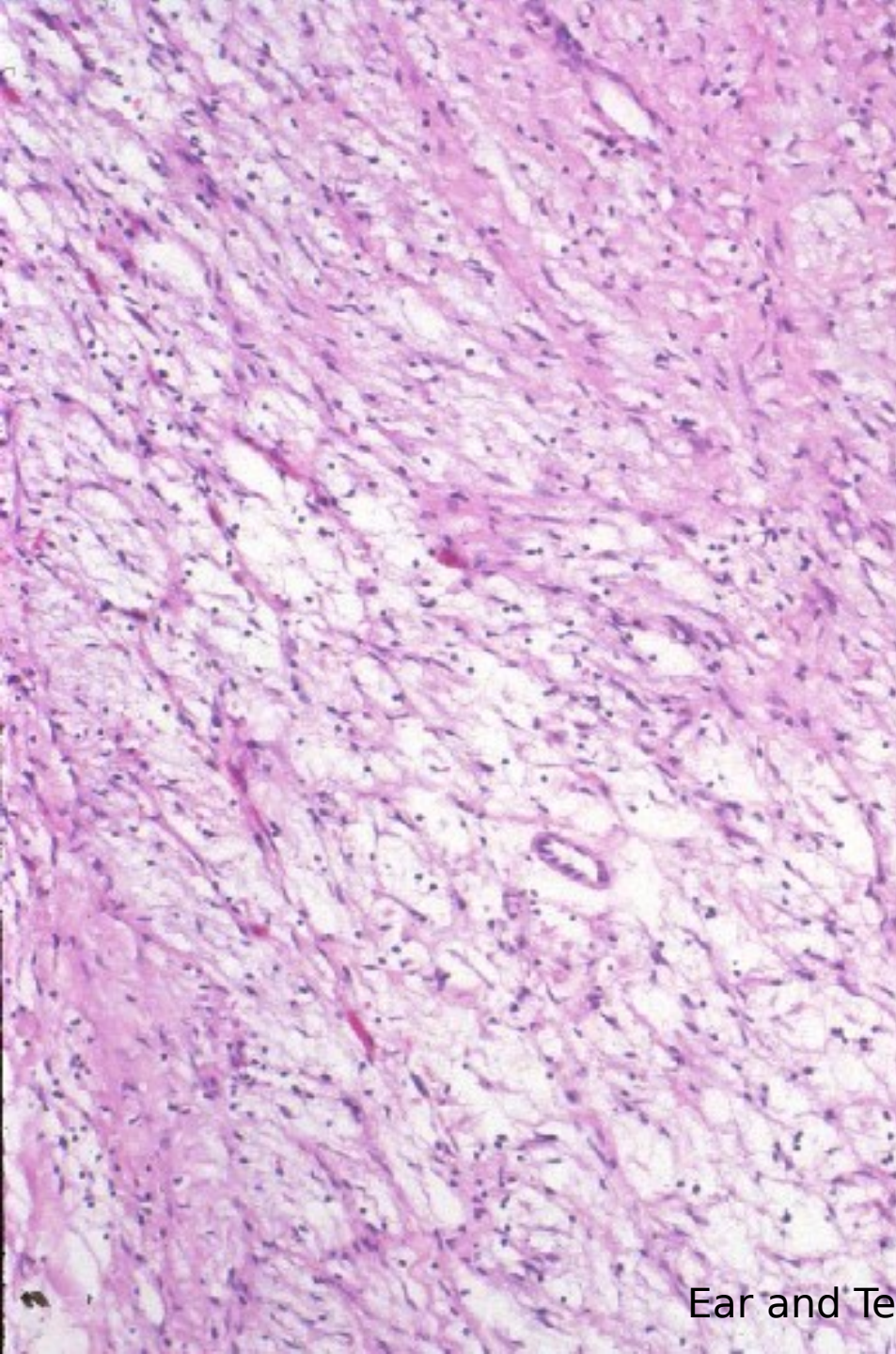


Case 5: A 36 year old female has headaches and hearing loss (unilateral). A cerebellopontine angle mass is identified.

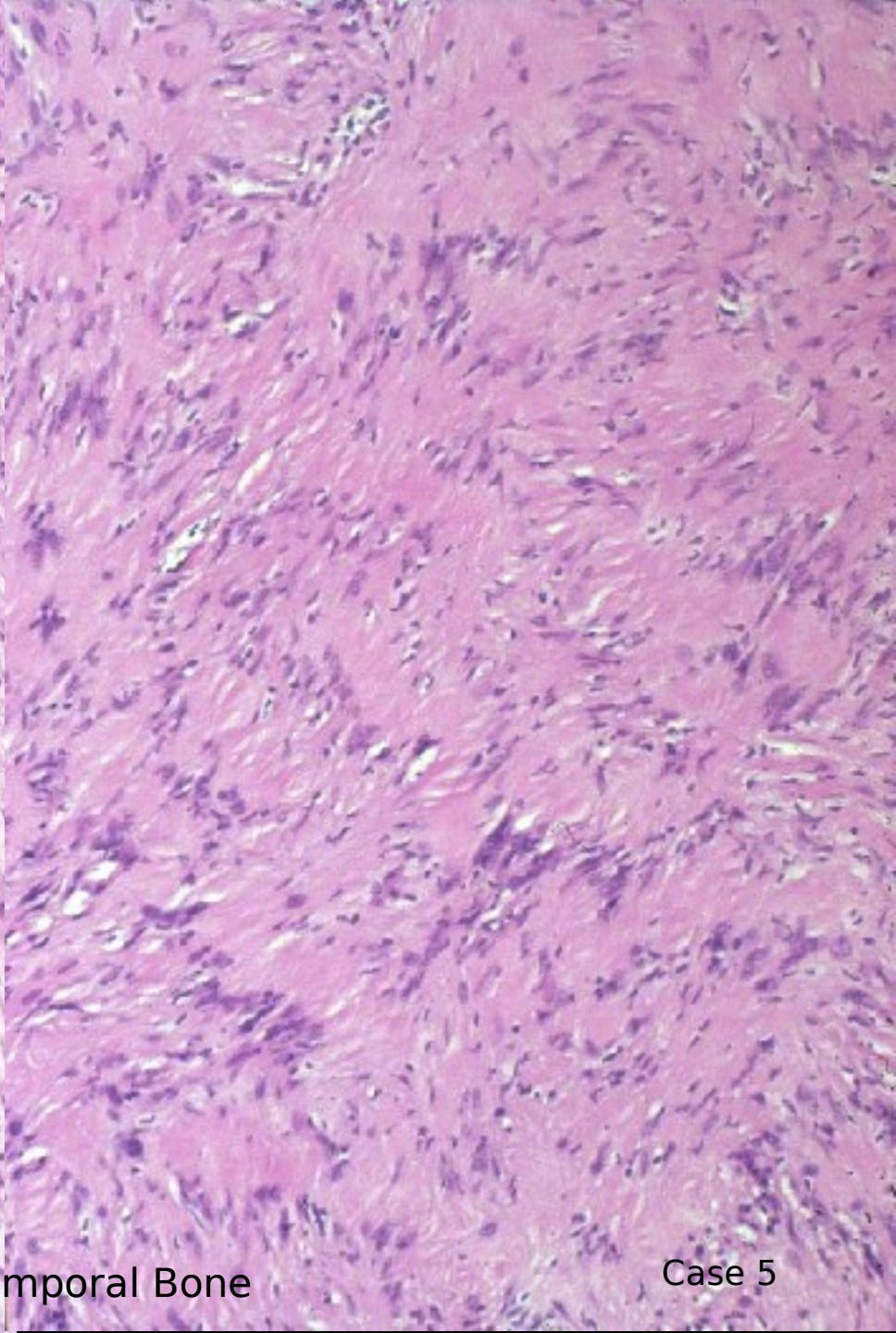
This cerebellopontine angle tumor :

1. Arises from the meninges.
2. Is of glial origin.
3. Shows only scattered S100 positive cells.
4. May be sporadic or may be associated with neurofibromatosis type 1.

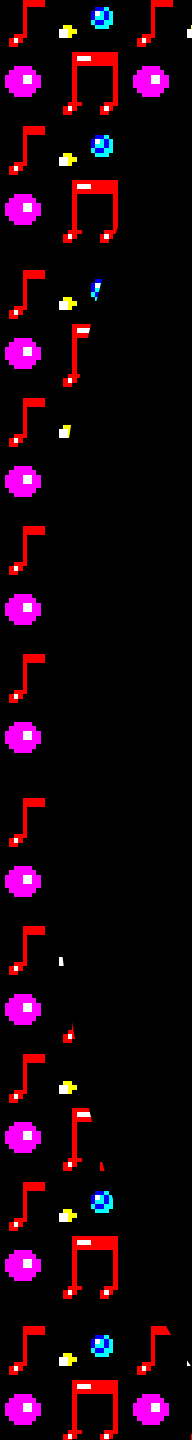




Ear and Temporal Bone



Case 5



Case 5: A 36 year old female has headaches and hearing loss (unilateral). A cerebellopontine angle mass is identified.

This cerebellopontine angle tumor :

1. Arises from the meninges.
2. Is of glial origin.
3. Shows only scattered S100 positive cells.
4. May be sporadic or may be associated with neurofibromatosis type 1.

Answer: D

Case 5: NEURILEMMOMA (ACOUSTIC NEUROMA)

Neurilemmomas are one of the common CP angle tumors (along with paraganglioma and meningioma). They are diffusely and strongly S100 positive, unlike neurofibromas. Compact cellular Antoni A areas alternate with hypocellular myxoid Antoni B areas. Nuclear palisading is seen in the Antoni A areas.

